Introduction

Advances in cardiac surgery and perioperative care in the past several decades have meant that over 85% of infants born with congenital heart disease (CHD) are now expected to reach adulthood. There are, in fact, relatively few conditions for which the surgical repair is completely and uniformly totally curative for the entire population. Cure requires that normal cardiovascular function be achieved and maintained, life expectancy is normal, and further medical evaluation for CHD is not required. It is estimated that there are currently over 500,000 adults in the USA with CHD, 55% of whom remain at moderate to high risk, and over 115,000 of whom have truly complex disease. Put another way, “the number of adults with CHD now equals the number of children with CHD.”

These patients bring with them problems related to complex postoperative anatomy and physiology which will not be familiar to physicians used to caring for adults, and also medical problems that accrue with aging, which will not be familiar to physicians used to caring for children. This problem has resulted in two American College of Cardiology sponsored Bethesda Conferences in the past several years, most recently in 2001. These panels have recommended the establishment of regionalized adult congenital heart centers which consist of a full coterie of professionals educated and experienced in the care of the adult with CHD. A specific recommendation was that non-cardiac surgery on CHD patients with moderate to complex disease be performed at an adult CHD center with the consultation of an anesthesiologist experienced with CHD.

This chapter reviews the organ system sequelae of longstanding CHD, both non-cardiac and cardiac; and the anatomy, pathophysiology, and surgical approach to the common lesions. Finally, perioperative and anesthetic outcomes are reviewed, and specific recommendations made for the anesthetic approach to adults with CHD.

Non-cardiac sequelae of longstanding congenital heart disease

Pulmonary sequelae

Lesions resulting in increased pulmonary blood flow or in obstruction to free pulmonary venous drainage can both cause increased interstitial fluid with decreased pulmonary compliance and increased work of breathing. Patients with cyanotic disease and chronic hypoxemia have increased minute ventilation with normal PaCO₂. Cyanotic patients appear to have a normal ventilatory response to hypercarbia but a blunted response to hypoxemia which resolves after surgical correction. End-tidal Pco₂ underestimates PaCO₂ in cyanotic patients with decreased, normal, or increased pulmonary blood flow.

Although enlarged, hypertensive pulmonary arteries or an enlarged left atrium can on occasion entrap the pulmonary arterial tree causing atelectasis, pneumonia, or focal emphysema in children; this is rare in adults. Hemoptysis is a finding of late stage Eisenmenger physiology, and thrombosis of upper lobe pulmonary arteries can occur in patients with Eisenmenger physiology and erythrocytosis. Prior thoracic surgery may have resulted in phrenic nerve damage.

The incidence of scoliosis in CHD patients is as high as 19%, is more common in children with cyanotic CHD, and may develop in adolescence, years after surgical correction of cyanosis. The interaction of cyanosis and early lateral thoracotomy in the development of scoliosis remains unclear. Although rare, scoliosis can be severe enough to impact pulmonary function.

The most serious complication of longstanding pulmonary hyperemia is the development of Eisenmenger physiology (see below). The age at which this develops depends on the underlying physiology (earlier at high altitude, for example), and also the level of the shunt. Patients with atrial level
shunts may not develop evidence of pulmonary vascular disease until late middle age.

**Hematologic sequelae**

Hematologic sequelae are predominantly a consequence of longstanding cyanotic CHD and include abnormalities of both red cell regulation and hemostasis. Chronic hypoxemia results in increased renal erythropoietin production. The relationship among oxygen saturation, 2,3-diphosphoglycerate, and red cell mass is relatively poor. The oxygen–hemoglobin dissociation curve is usually normal or minimally right-shifted. Most patients establish an equilibrium state. They have a stable hematocrit and are iron replete. Some patients, however, develop excessive hematocrits and are iron deficient, resulting in a hyperviscous state. Symptoms of hyperviscosity are rare at hematocrits less than 65% if the patient is not iron deficient (Table 13.1). Iron deficient red cells are less deformable than iron replete red cells, and will cause increased viscosity for the same hematocrit. Iron deficiency can be related to inappropriate, repeated phlebotomies in an attempt to reduce hematocrit. The blood cells will be microcytic and hypochromic in the face of erythrocytosis. Treatment with oral iron should be undertaken with care, as rapid increases in hematocrit can ensue.

Erythrocytosis and hyperviscosity are associated with the development of cerebral venous thrombosis in children less than 4 years old, but not in adults with cyanotic CHD, regardless of hematocrit. Therapy is recommended for temporary relief of symptomatic hyperviscosity only (not due to dehydration). Symptoms usually regress within 24 h of a partial isovolumic exchange transfusion. It is rare for adult patients to require removal of more than 1 U of blood. Phlebitomy should only be undertaken in symptomatic patients, and not to treat asymptomatic high hematocrits. Phlebotomized blood can be banked for autologous perioperative transfusion if needed. Prolonged preoperative fasting should be specifically avoided in these patients, as rapid increases in hematocrit can accompany dehydration. Each volume of erythrocytotic blood contains less plasma than normal blood (Fig. 13.1).

A variety of hemostatic abnormalities have been described in cyanotic patients. Bleeding diatheses are uncommon at hematocrits less than 65%, although surgical bleeding can occur. Generally the degree of the bleeding diathesis mirrors the hematocrit. Platelet counts are typically low normal, and are occasionally low, but bleeding is not related to the thrombocytopenia. When corrected for the decreased plasma volume in each blood sample, total plasma platelet count is closer to normal (Fig. 13.1). Abnormalities of platelet function have also been reported. Patients with synthetic vascular anastamoses or low-pressure conduits are often maintained on chronic antiplatelet therapy.

Abnormalities of both the intrinsic and extrinsic coagulation pathways with abnormalities of a variety of specific clotting factors have been inconsistently described in cyanotic patients. Fibrinolytic pathways are normal. On occasion patients with both cyanotic and acyanotic CHD have been described with deficiencies of the largest von Willebrand factor multimers, which have corrected following corrective cardiac surgery.

The decreased plasma volume in erythrocytotic blood may cause spurious results of the prothrombin and partial thromboplastin times. The fixed amount of anticoagulant in the sample tube presumes a normal plasma volume and may be excessive for an erythrocytotic sample (Fig. 13.1). If informed of the patient’s hematocrit, the clinical laboratory can provide an appropriate tube. Correcting to an idealized hematocrit of 45%, the appropriate amount of citrate can be added to the tube as follows:

\[ \text{Milliliter citrate} = (0.1 \times \text{blood volume collected}) \times [100 - \text{patient’s hematocrit}/55]. \]

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Table 13.1 Signs and symptoms of hyperviscosity syndrome.

<table>
<thead>
<tr>
<th>Sign/Symptom</th>
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</thead>
<tbody>
<tr>
<td>Headache</td>
</tr>
<tr>
<td>Faintness, dizziness, light headedness</td>
</tr>
<tr>
<td>Blurred or double vision</td>
</tr>
<tr>
<td>Fatigue</td>
</tr>
<tr>
<td>Myalgias, muscle weakness</td>
</tr>
<tr>
<td>Paresthesias of fingers, toes, or lips</td>
</tr>
<tr>
<td>Depressed mentation, a feeling of dissociation</td>
</tr>
</tbody>
</table>

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Fig. 13.1 The effect of a fixed amount of citrate anticoagulant in tubes of blood with normal and increased hematocrit. The fixed anticoagulant volume combined with decreased plasma volume in the erythrocytotic blood results in artifactual elevation of the prothrombin and partial thromboplastin times. Similarly, although the concentrations of platelets are identical in both samples of plasma, the platelet count per milliliter of whole blood (which is reported by the laboratory) will be lower in erythrocytotic blood. Reproduced with permission from Baum VC. The adult with congenital heart disease. *J Cardiothorac Vasc Anesth* 1996; 10: 261–82.
Due to the excessive hemoglobin turnover in cyanotic CHD, adult patients with cyanotic CHD have an increased incidence of calcium bilirubinate gallstones, and biliary colic can develop years after cardiac surgery has resolved the cyanosis. Factors besides intrinsic hemostatic defects can increase the risk of excessive perioperative bleeding in patients with cyanotic CHD, particularly during thoracic surgery. These include increased tissue vascularity, elevated systemic venous pressure, and abnormal aortopulmonary and transpleural collateral vessels. In addition, many patients will have had prior intrathoracic surgery.

Renal sequelae

Abnormal renal histopathology with chronic cyanotic CHD includes hypercellular glomeruli with basement membrane thickening, focal interstitial fibrosis, tubular atrophy, and hyalination of afferent and efferent arterioles. High plasma uric acid levels can be found in adults with cyanotic CHD. Although one might presume that this is from increased urate production, it is rather due to inappropriately low fractional uric acid excretion. This enhanced urate reabsorption is believed due to renal hypoperfusion with a high filtration fraction. Urate stones and urate nephropathy are, however, rare. Arthralgias are common, but gouty arthritis is less frequent than would be expected from the degree of hyperuricemia.

Neurologic sequelae

Adults with unmodified or persistent intracardiac shunts remain at risk for paradoxical emboli. Even patients with a predominant left-to-right shunt are at some risk. Although it has been said that unlike children, adults with cyanotic CHD are not at risk for the development of cerebral thrombosis no matter the level of hematocrit, that conclusion has been challenged. In any event, adults do remain at risk for the development of brain abscesses. A healed childhood brain abscess can serve as a nidus for seizures throughout adulthood.

Prior thoracic surgery can have caused iatrogenic peripheral nerve damage. Surgery at the apices of the lung, such as Blalock–Taussig shunts, patent ductus arteriosus (PDA) ligation, pulmonary arterial band, and coarctation repair in particular, are associated with injury to the recurrent laryngeal nerve, the phrenic nerve, and the sympathetic chain. Resultant injuries can be permanent.

Vascular access considerations

Both congenital abnormalities and alterations due to cardiac catheterization or surgery can affect the suitability of a variety of vessels for cannulation by the anesthesiologist. These are summarized in Table 13.2.

Table 13.2 Vascular access considerations.

<table>
<thead>
<tr>
<th>Vessel</th>
<th>Potential problem</th>
</tr>
</thead>
<tbody>
<tr>
<td>Femoral vein(s)</td>
<td>In older patients may have been ligated if cardiac catheterization done by cutdown. In younger patients may be thrombosed after use of larger therapeutic catheters</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>Some lesions (particularly asplenia) associated with discontinuity of the inferior vena cava. Will not be able to pass a catheter from the groin to the right atrium</td>
</tr>
<tr>
<td>Left subclavian and pedal arteries</td>
<td>Blood pressure will be low in the presence of coarctation of the aorta or following subclavian flap repair (subclavian artery only), and variably so if postoperative recoarctation</td>
</tr>
<tr>
<td>Subclavian artery</td>
<td>Blood pressure low with classic Blalock–Taussig shunt on that side, and variably so with modified Blalock–Taussig</td>
</tr>
<tr>
<td>Right subclavian artery</td>
<td>Blood pressure artificially high with supravalvar aortic stenosis (Coanda effect)</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>Risk of catheter-related thrombosis with Glenn operation</td>
</tr>
</tbody>
</table>

Pregnancy

As more children grow into adulthood with CHD, so will more of them become pregnant. The physiologic changes of pregnancy, labor, and delivery can significantly alter the physiologic status of these patients. Readers are referred to more specialized books for a more complete discussion of the pregnant woman with CHD. Pregnancy considerations are included under specific defects listed below.

Pregnancy can be carried successfully to term with vaginal delivery for women with most congenital cardiac lesions. Pulmonary hypertension, depressed ventricular function, and cyanosis are predictors of maternal and fetal complications. Patients with Eisenmenger physiology are at particularly high risk. Up to 47% of cyanotic women will develop deterioration of functional capacity during pregnancy.
Anticoagulation is recommended for cyanotic women and women with Eisenmenger physiology during the third trimester and the first postpartum month. The decrease in systemic vascular resistance that accompanies pregnancy is better tolerated in patients with regurgitant lesions.

Bacterial endocarditis prophylaxis is not currently recommended for uncomplicated deliveries, although it is probably common clinical practice. Episiotomies would be an indication for prophylaxis.

**Psychosocial issues**

Adolescents often have psychological peculiarities well known to pediatricians, and teenagers with CHD are no different. Issues of denial, sense of immortality, and desire for risk taking can all impact on optimally caring for these adolescents as they transition into adulthood. Body conscious adolescents may struggle with bodies that are scarred due to prior surgery and may have physical limitations. Although most adolescents and adults with CHD function quite well, adults with CHD are less likely to be married or cohabiting, and are more likely to be living with their parents.24

Adult CHD patients may have difficulty in obtaining life and health insurance after they are no longer covered under their parents’ policies.25 Life insurance is somewhat more readily available than in the past, but policies vary widely among insurers.26

**Cardiac sequelae**

The hemodynamic effects of an anatomical cardiac defect can be compounded by time, and modified by imposed chronic cyanosis or pulmonary vascular disease. Myocardial dysfunction can be inherent to the CHD, but can also be due to surgical injury, including inadequate intraoperative myocardial protection.27,28 This is particularly true of now middle-aged adults who had surgical repairs several decades ago. Although the basic pathophysiology might be well understood by those caring for children with CHD, the natural history of these lesions may be unexpected. Some patients with dysmorphic syndromes will develop heart disease in adult life. For example 46% of a young adult Down’s syndrome population without CHD developed mitral valve prolapse, and a small number developed aortic insufficiency.29 The large number of cardiac lesions and subtypes, compounded with an array of surgical palliative and corrective procedures, make a complete cataloging of all defects and modifications impossible in this context. This chapter is primarily devoted to the more common and physiologically important defects. Both short-term and long-term surgical results from older series may not reflect current results.

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**Acyanotic lesions**

**Atrial septal defect and partial anomalous pulmonary venous connection**

Both the natural history and the outcome after surgery for partial anomalous pulmonary venous return are similar to that of the physiologically similar secundum atrial septal defect (ASD).30–32 Because patients with otherwise uncomplicated ASDs often remain asymptomatic until adulthood, ASDs account for about one-third of CHD discovered in adults. Survival of unrepaired defects into adulthood is routine, but complications developing in adulthood provide the rationale for routine childhood correction. There is a mortality of 6%/year over 40 years of age,30–32 and essentially all patients over 60 years of age are asymptomatic. Patients with large, unrepaired defects often die of right ventricular failure or atrial tachyarrhythmias in their thirties or forties.33 In addition to atrial tachyarrhythmias and paradoxical emboli, left-to-right shunting through the defect can increase with aging. Systemic hypertension and/or ischemic coronary disease can occur with aging, and both decrease left ventricular diastolic compliance, which increases the left-to-right shunt. After the age of 40 years, patients can develop pulmonary vascular disease, now pressure loading the chronically volume loaded right ventricle. Mitral insufficiency can develop in adulthood and is significant in about 15% of adult patients.34

Incomplete resolution of right ventricular dilation has been reported with surgical closure after 5 years of age.35 Left ventricular dysfunction has been reported by some in patients having surgical closure in adulthood.36 Postoperative survival in patients without pulmonary vascular disease is similar if operated on before 24 years of age, but survival is worse if surgery is done between 25 and 41 years of age, and worse yet after 41 years of age.37

Pregnancy is uncomplicated in the vast majority but, with the hypervolemia of pregnancy, heart failure can develop during pregnancy with larger defects. Peripheral venous thrombosis carries with it the risk of paradoxical embolization.

**Ventricular septal defect**

The long-term natural history of ventricular septal defect (VSD) has been reviewed in detail.38 More than 75% of small and even moderate-sized VSDs close spontaneously during childhood by a gradual ingrowth of surrounding septum. Over 90% of those defects that will close spontaneously will have closed by 10 years of age. Other mechanisms of natural closure include closure by tricuspid valve tissue, prolapsed aortic valve tissue, and endocarditis. There is an incidence of aortic insufficiency in adults with VSD from prolapse into the defect.39 Otherwise, a small VSD in the adult is of no hemodynamic import, other than the continuing risk of endocarditis.
Pulmonary vascular disease can progress if closure of a large VSD is delayed. Several studies have shown possible ventricular dysfunction years after surgical closure. However, most of these patients had surgery late, by current standards. It appears that the changes of chronic volume overload resolve if surgical correction is undertaken by 5 years of age. Pregnancy is well tolerated in the absence of pulmonary hypertension or pre-existing heart failure. Pregnancy with a spontaneous or surgically closed defect carries no additional risk, in the absence of additional cardiac problems.

**Patent ductus arteriosus**

Patent ductus arteriosus rarely close spontaneously after the neonatal period. In addition to the consequences of chronic left-to-right shunting, in the adult the PDA may become calcified or aneurysmally dilated with the risk of rupture. This increases the risk of surgery, which will rarely require the use of cardiopulmonary bypass. Unrepaired, one-third of patients die of heart failure, pulmonary arterial hypertension or endocarditis by 40 years of age, and two-thirds by the age of 60 years. Small PDAs do not carry a hemodynamic risk for pregnancy.

**Coarctation of the aorta**

There is a significant morbidity and mortality from unoperated coarctation of the aorta in the adult. There is a 25% mortality by 20 years of age, 50% by 30 years of age, 75% by 50 years of age, and 90% by 60 years of age. Causes of death include left ventricular failure, rupture of cerebral aneurysms, and dissection of a post-coarctation aneurysm. Left ventricular failure can develop in unrepaired adults over 40 years of age. Unless repair is undertaken early in life, there is an incremental risk for the development of premature coronary atherosclerotic disease. Even with operation, coronary artery disease is the leading cause of death 11–25 years after operation. Bicuspid aortic valve is a common coexisting lesion, and often does not become stenotic until middle age or older, although it is always an endocarditis risk. Coarctation can also be associated with functionally significant mitral valve abnormalities. Patients who have had coarctation repairs in childhood may develop aneurysms at the site of the repair, or restenosis of the repaired area in adolescence or adulthood. Half of patients operated on after 40 years of age have persistent hypertension, and many of the remainder will have abnormal hypertensive responses to exercise. Long-term survival after surgery is worse the older the patient at the time of surgery, with a 15-year survival of only 50% in patients having surgery at over 40 years of age.

Hypertension can be exacerbated during pregnancy in women with unoperated coarctation, with the risks of aortic dissection or rupture, heart failure, angina, and rupture of a circle of Willis aneurysm. Blood pressure control is of great importance during pregnancy. Most aortic ruptures during pregnancy occur prior to labor and delivery. Epidural analgesia would help minimize hypertension during delivery.

**Aortic stenosis**

Most adult patients with aortic stenosis have a bicuspid aortic valve. Although endocarditis risk is lifelong, symptoms often do not develop until late middle age or later. Once symptoms develop (angina, syncope or near syncope, heart failure) survival is markedly shortened: median survival is 5 years after the development of angina, 3 years after syncope, and 2 years after heart failure. Most mothers with aortic stenosis can have safe pregnancies with vaginal deliveries. Severe aortic stenosis (valve area < 1.0 cm²) may cause maternal clinical deterioration and significant maternal and fetal mortality. Hemodynamic monitoring during delivery is critical with maintenance of preload and avoidance of vasodilation and hypotension. When required, percutaneous balloon valvuloplasty appears to carry lower risk than open valvotomy during pregnancy.

**Pulmonary valve stenosis**

Apart from neonates with critical pulmonic stenosis, long-term asymptomatic survival is routine. Mild pulmonic stenosis in the adult does not require surgical correction; there is a 94% survival 20 years after diagnosis. However with aging, right ventricular fibrosis and right ventricular failure can develop, which is the most common cause of death, occurring usually in the fourth decade. Essentially all patients who have relief of stenosis surgically or by balloon valvuloplasty have normal postoperative right ventricular function, but abnormal ventricular function may not completely normalize after late correction. Isolated pulmonary stenosis even of a severe degree, is usually well tolerated during pregnancy, despite the volume overload.

**Congenitally corrected transposition of the great vessels (l-transposition, ventricular inversion)**

Most patients whose anatomic right ventricle is the systemic ventricle as an isolated defect will have normal biventricular function through early adulthood, but can develop right ventricular failure with increasing age. Second or third degree heart block occurs with an incidence of about 2%/year, and more than 75% of patients have some degree of heart block, although the intrinsic pacemaker remains above the bundle of His with a narrow QRS. l-Transposition is associated with an Ebstein-like deformity of the tricuspid valve (in the systemic ventricle). There is a significant incidence of tricuspid insufficiency (physiologically analogous to mitral insufficiency in the normal heart) even in patients without this...
Ebstein-like malformation, and the incidence is higher still in patients with this valve deformity.53

Ebstein’s anomaly of the tricuspid valve
Following tricuspid valve replacement (the current approach is repair if possible) up to 25% of patients will have high-grade atrioventricular block. There is often associated a right-sided bypass tract resulting in Wolff–Parkinson–White accelerated conduction, allowing rapid ventricular rates and possible development of ventricular fibrillation. This is a particular concern as 25–30% of patients will develop supraventricular tachyarrhythmias in addition to the fraction that will develop atrial fibrillation as a consequence of aging.

In the absence of marked cyanosis, pregnancy and delivery are generally well tolerated, but with an increased incidence of prematurity and fetal loss.

Cyanotic lesions

Tetralogy of Fallot
Tetralogy of Fallot is the most common cyanotic lesion encountered in adults. Unoperated, approximately 25% of patients will survive to adolescence, following which the mortality is 6.6%/year. Only 3% will survive to the age of 40 years.54 Unlike children, adolescents and adults with tetralogy do not develop hypercyanotic “tet spells.” The outcome in patients surgically corrected as adults is worse compared to surgical correction in childhood.55

Although the VSD component is currently approached through the right atrium, adult patients may have had repair via a right ventriculotomy. Right ventricular function in these patients can have an abnormal response to exercise. Repair at an earlier age (<12 years of age) results in better long-term right ventricular function. In the (now uncommon) un repaired adult patient, the development of systemic hypertension in adult life will impose an additional load on both ventricles, not just the left ventricle. The increased systemic resistance can decrease the right-to-left shunt and improve cyanosis, but at the expense of right- or biventricular failure.

Up to 5.5% of postoperative patients may have sudden death or require treatment for ventricular tachycardia, often years after surgical correction.56 The foci for these arrhythmias are typically in the right ventricular outflow tract and can be ablated. However, premature ventricular contractions and even non-sustained ventricular tachycardia are not uncommon but may not be associated with sudden death,57 making it difficult to know which patients to treat. Additional long-term complications include chronic pulmonary insufficiency and aneurysm formation at a right ventricular outflow tract patch.

Women who have had a good surgical correction without residual defects should tolerate pregnancy and delivery well. Women with uncorrected tetralogy of Fallot, particularly those with significant cyanosis, have a high incidence of fetal loss (80% with hematocrit > 65%). The fall in systemic resistance with pregnancy and delivery can worsen cyanosis, and the physiologic volume load can exaggerate failure of both ventricles.

Transposition of the great arteries (d-transposition)
With a 1-year mortality of approximately 100%, all adults with d-transposition will have had some type of surgical correction. Many adults will have had atrial type repairs, of either the Mustard or Senning type. Some teenagers will be young enough to have had repair by an arterial switch operation. Some adults will have had repair of d-transposition and VSD with a Rastelli type repair. Atrial repairs result in a systemic right ventricle. Patients who have had an atrial type repair have consistently abnormal right ventricular function, with a right ventricular ejection fraction of about 40%. It has been suggested that the earlier the surgery the better the right ventricular function, although it remains abnormal.58 Right ventricular dysfunction can be progressive.59

There is a significant incidence of late electrophysiologic sequelae after atrial repair, including sinus node dysfunction (bradycardia), junctional escape rhythms, atrioventricular block, and supraventricular tachyarrhythmias. These atrial arrhythmias can result in sudden death, presumably from 1 : 1 conduction causing ventricular fibrillation.60 The frequency of tachyarrhythmias increases after the tenth postoperative year.

It is still too premature to know the very long-term outcome after the arterial switch operation. Many of these children have abnormal resting myocardial perfusion, and the implication for the development of coronary artery disease in adulthood remains unknown.

Pregnancy and delivery are generally well tolerated after an atrial or Rastelli repair, however right ventricular failure and worsening functional capacity can occur. There is an increased incidence of prematurity and small infants in the offspring of these women.

Single ventricle anatomy
This large rubric includes such lesions as tricuspid atresia and more complex anatomy with a single ventricle, and thus long-term survival depends on the type and degree of coexisting cardiac malformations. Both pulmonary stenosis (protecting the pulmonary vasculature from excessive flow) and a competent atrioventricular valve improve long-term survival. A single ventricle of left ventricular morphology
allows for better ventricular function than does one of the right ventricular type.\textsuperscript{61} Palliation with an aortopulmonary shunt is associated with volume loading of the single ventricle and decreasing function with age.

Pregnancies have been reported in women with single ventricles. There is a high incidence of fetal loss, premature delivery, and small infants. Stable patients, however, can have vaginal delivery. The risk of pregnancy increases significantly with increasing degrees of cyanosis.

**Fontan physiology**

The Fontan operation has undergone many iterations and modifications since its original report in 1971. There is evidence that despite early improvement in function, these patients, at least those done early in the experience with this operation, have continued decline in function with continued long-term mortality.\textsuperscript{62}

**Truncus arteriosus**

Essentially all patients who survive to adolescence will have had surgical repair. Although conduits placed in early childhood will be outgrown, valved conduits placed in late childhood should suffice for adult size. There can be ongoing problems with incompetence or stenosis of both the truncal valve, now analogous to the aortic valve, and the valved right ventricle to pulmonary artery conduit. Because the conduit often lies immediately behind and in close proximity to the sternum, it can be at very high risk of accidental incision during later sternotomy.

**Eisenmenger’s syndrome**

Eisenmenger physiology is compatible with survival into adulthood.\textsuperscript{63} Survival is 80\% 10 years after diagnosis, and 42\% at 25 years.\textsuperscript{64} Significant mortality occurs with non-cardiac surgery and pregnancy.\textsuperscript{65} The onset of irreversible pulmonary vascular disease depends on the degree of shear rate, and for atrial level shunts such as ASDs it may not develop until mid-life. Patients with pulmonary vascular disease face significant potential perioperative risks and constitute a major proportion of adults referred for anesthetic evaluation prior to non-cardiac surgery. Findings are summarized in Table 13.3.

Fixed pulmonary vascular resistance precludes rapid adaptation to intraoperative hemodynamic changes. Changes in systemic vascular resistance are mirrored by changes in the degree of right-to-left shunting. Systemic vasodilators, including regional anesthesia, must be used with caution, and close assessment of intravascular volume is important. Epidural anesthesia has been used successfully in these patients, but the local anesthetic should be delivered in small increments.\textsuperscript{66} Postoperative postural hypotension can increase the degree of right-to-left shunting, and these patients should be cautioned to change position slowly.

Placement of pulmonary artery catheters is problematic and not without potential complication in patients with pulmonary vascular disease, who can also have hemostatic defects associated with erythrocytosis.\textsuperscript{67} Pulmonary hypertension is a risk factor for pulmonary artery rupture. Right-to-left intracardiac shunting and abnormal cardiac anatomy may make passage to the pulmonary artery difficult without fluoroscopy. Given that the relative resistances of the systemic and pulmonary beds will be reflected in systemic oxygen saturation, which is readily measured by pulse oximetry, and measurements of thermodilution output will not accurately reflect systemic output, the value of a pulmonary artery catheter in these patients is minimal at best and they are essentially never indicated. One possible exception is the patient with pulmonary vascular disease and an ASD who is at risk to develop right ventricular failure if suprasystemic right ventricular pressures develop.\textsuperscript{68}

Fixed pulmonary vascular resistance is by definition unresponsive to pharmacologic manipulation. Nevertheless it would seem prudent to avoid those factors known to exacerbate pulmonary resistance including hypothermia, hypercarbia, acidosis, hypoxia, and α-adrenergic agonists. Although the last of those is commonly listed, it has seemed that in the context of pulmonary vascular disease due to a shunt lesion, systemic vasoconstrictive effect predominates and systemic oxygen saturation will increase.

Appropriate nerve blocks offer an attractive alternative to general anesthesia. If patients undergo general anesthesia, consideration should be given to returning them to an intensive care unit for gradual emergence and close observation. Because of the increased perioperative risk, patients should be observed at least overnight in an intensive care type of unit, particularly if they have not had any recent surgery or

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**Table 13.3 Signs, symptoms, and findings with Eisenmenger’s syndrome.**

- Physical examination: Loud pulmonic component of the second heart sound, single or narrowly split second heart sound, Graham-Steell murmur of pulmonary insufficiency, pulmonic ejection sound (“click”)
- Chest radiography: Decreased peripheral pulmonary arterial markings with prominent central pulmonary vessels (“pruning”)
- Electrocardiogram: Right ventricular hypertrophy
- Impaired exercise tolerance
- Exertional dyspnea
- Palpitations (often due to atrial fibrillation or flutter)
- Complications from erythrocytosis/hyperviscosity (see text)
- Hemoptysis from pulmonary infarction or rupture of pulmonary vessels or aortopulmonary collateral vessels
- Complications from paradoxical embolization
- Syncope from inadequate cardiac output or arrhythmias
- Heart failure (usually end-stage)
anesthesia and their response will be unknown. Ambulatory surgery is possible, however, for patients having had uncomplicated minor surgical procedures with sedation or nerve block.

Pregnancy carries with it a very high mortality risk—30% of all pregnancies end in maternal death, and a successful first pregnancy does not preclude maternal death during a subsequent pregnancy. The changes in hemodynamics of both pregnancy and delivery increase maternal risk. Pulmonary embolism (macro and micro) has caused peripartum deaths, and death can occur days after delivery. Women should be carefully monitored, with arterial catheters, during delivery. Epidural analgesia, delivered slowly and carefully, can mitigate many of the deleterious hemodynamic changes of active labor. There is a high incidence of premature deliveries. Pulmonary hypertension and pregnancy has been reviewed in detail.

### Perioperative and anesthetic outcome

In a recent retrospective review from Texas Children’s Hospital (TCH), the anesthetic management and immediate outcome of adult and teenaged patients undergoing surgery for CHD were compared to lesion-matched control patients under 6 years of age. The primary outcome variable was death within 30 days of surgery, and secondary outcomes were major neurological morbidity, mechanical ventilation beyond 24 h postoperatively, and length of ICU and hospital stay.

Perioperative and outcome data from this review are presented in Table 13.4. All patients who experienced major neurological morbidity or perioperative mortality were in the older patient group, and all of those were undergoing repeat operations. Four patients died within 30 days of surgery, none in the operating room. No deaths or other major intraoperative events occurred in any patient undergoing a first-time operation, whether in the younger or older patient group.

The anesthetic agents used for both younger and older patients were very similar. Etomidate was used more frequently for induction of anesthesia in older patients, and sevoflurane was used more commonly in younger patients. All patients received a narcotic-based technique using fentanyl. Also there was no difference between groups regarding the use of single or multiple inotropic agents.

Fifty-nine percent of the older patients vs. 15% of younger patients required antiarrhythmic treatment, and greater numbers of older patients received lidocaine, amiodarone, and magnesium sulfate. Temporary cardiac pacing was used, and defibrillation performed more frequently in the older patients. Of the arrhythmias requiring treatment, 58% were ventricular in the adults compared to 24% in the control patients.

The only other published study regarding perioperative outcome of adults with CHD is from the Royal Brompton Hospital (RBH). These authors report a slightly greater overall mortality, 6.8% compared to 4.7% in the TCH study. They also found reoperation to be a significant risk factor for early postoperative mortality, and that the number of previous operations correlated with increased mortality. Cyanosis and increasing age were also correlated with increased mortality. Compared to the TCH study, patients in the RBH study were significantly older, with a mean age of 31 years, and one-third of the non-survivors in the RBH study were over 50 years of age.

From these retrospective reviews, there appears to be an increased incidence of perioperative morbidity among older patients with CHD undergoing cardiac surgery, and certain groups of patients have the greatest risk, particularly the single ventricle patient. Because there is improved outcome from the Fontan procedure, the number of these patients requiring surgery in adulthood will increase in the future. In these patients, the transition from spontaneous to positive pressure ventilation will decrease pulmonary blood flow and cardiac output. Cardiac output is also significantly compromised by non-sinus rhythm, hypovolemia, or myocardial depressant anesthetics. The only patient requiring epinephrine after the induction of anesthesia in the TCH review was scheduled for a Fontan revision.

Another group of patients with greater risk of mortality or major morbidity are those with cyanosis, especially those requiring repeat sternotomy. These two factors were the best predictors of early mortality in the RBH study, and all deaths and major complications occurred among these patients in the TCH study. There are several reasons for this observation. First, longstanding cyanosis leads to increased risk for coagulopathy and organ dysfunction. Second, most of these patients had ventricular dysfunction, which renders the myocardium more vulnerable to the ischemic insult from cardiopulmonary bypass and aortic cross-clamping, thereby increasing the possibility of postoperative ventricular failure and arrhythmias.

### Anesthetic management

Based on the above data and our experience, we recommend the following management for adult patients undergoing congenital heart surgery:

#### Preoperative preparations:

1. Patient data should be presented to a multidisciplinary group consisting of cardiologists, surgeons, and anesthesiologists. Data analysis includes: laboratory results, cardiac catheterization, echocardiography, Holter monitor results, chest radiograph, and magnetic resonance imaging. Among this group of specialists a consensus can be developed regarding the timing of surgery and surgical options, which may include cardiac transplantation.
PART 3 Preoperative considerations

<table>
<thead>
<tr>
<th></th>
<th>Older patients</th>
<th>Control patients</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient data</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patient Number</td>
<td>85</td>
<td>170</td>
<td></td>
</tr>
<tr>
<td>Age (yrs) (mean ± SD)</td>
<td>21.2 ± 10</td>
<td>2.0 ± 1.5</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Age range (yrs)</td>
<td>13–71</td>
<td>0–5</td>
<td></td>
</tr>
<tr>
<td>Weight (kg) (mean ± SD)</td>
<td>61.6 ± 22</td>
<td>10.7 ± 4.6</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Reoperation (no. (%))</td>
<td>49 (58)</td>
<td>51 (30)</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Cyanosis (no. (%))</td>
<td>19 (22)</td>
<td>90 (53)</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Surgery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fontan or revision (no. (%))</td>
<td>14 (16)</td>
<td>35 (21)</td>
<td>0.536</td>
</tr>
<tr>
<td>Conduit change or placement (no. (%))</td>
<td>9 (11)</td>
<td>15 (9)</td>
<td>0.820</td>
</tr>
<tr>
<td>Valve repair/replacement (no. (%))</td>
<td>10 (12)</td>
<td>18 (11)</td>
<td>0.944</td>
</tr>
<tr>
<td>ASD/VSD repair (no. (%))</td>
<td>18 (21)</td>
<td>34 (20)</td>
<td>0.956</td>
</tr>
<tr>
<td>Complex repair (no. (%))</td>
<td>28 (33)</td>
<td>63 (37)</td>
<td>0.611</td>
</tr>
<tr>
<td>Other (no. (%))</td>
<td>6 (7)</td>
<td>5 (3)</td>
<td>0.321</td>
</tr>
<tr>
<td>Intraoperative data (min)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anesthesia time (mean ± SD)</td>
<td>451 ± 149</td>
<td>383 ± 95</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Surgical time (mean ± SD)</td>
<td>349 ± 139</td>
<td>290 ± 91</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>CPB time (mean ± SD)</td>
<td>159 ± 85</td>
<td>140 ± 63</td>
<td>0.065</td>
</tr>
<tr>
<td>Aortic cross-clamp time (mean ± SD)</td>
<td>91 ± 51</td>
<td>84 ± 45</td>
<td>0.315</td>
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<tr>
<td>Temperature outcome</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lowest temp on bypass (°C) (mean ± SD)</td>
<td>27.9 ± 4.2</td>
<td>27.1 ± 3.8</td>
<td>0.135</td>
</tr>
<tr>
<td>Bleeding requiring transfusion (no. (%))</td>
<td>41 (48)</td>
<td>90 (53)</td>
<td>0.565</td>
</tr>
<tr>
<td>Dysrhythmia requiring treatment (no. (%))</td>
<td>43 (51)</td>
<td>26 (15)</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Inotropes (no. (%))</td>
<td>69 (81)</td>
<td>149 (88)</td>
<td>0.232</td>
</tr>
<tr>
<td>Vascular access problems (no. (%))</td>
<td>10 (12)</td>
<td>53 (31)</td>
<td>0.001</td>
</tr>
<tr>
<td>CPR (no. (%))</td>
<td>2 (2)</td>
<td>0</td>
<td>0.210</td>
</tr>
<tr>
<td>↓ BP on induction requiring epinephrine (no. (%))</td>
<td>1 (1)</td>
<td>0</td>
<td>0.420</td>
</tr>
<tr>
<td>Femoral bypass (no. (%))</td>
<td>2 (2)</td>
<td>0</td>
<td>0.210</td>
</tr>
<tr>
<td>Massive hemorrhage (no. (%))</td>
<td>4 (5)</td>
<td>0</td>
<td>0.021*</td>
</tr>
<tr>
<td>IABP/VAD/ECMO (no. (%))</td>
<td>0</td>
<td>0</td>
<td>1.000</td>
</tr>
<tr>
<td>Postoperative ventilation &gt; 24 h (no. (%))</td>
<td>17 (20)</td>
<td>54 (32)</td>
<td>0.068</td>
</tr>
<tr>
<td>Neurologic complication (no. (%))</td>
<td>3 (4)</td>
<td>0</td>
<td>0.065</td>
</tr>
<tr>
<td>Intraoperative death (no. (%))</td>
<td>0</td>
<td>0</td>
<td>1.000</td>
</tr>
<tr>
<td>Postoperative death (no. (%))</td>
<td>4 (5)</td>
<td>0</td>
<td>0.021*</td>
</tr>
<tr>
<td>Length of stay</td>
<td>ICU LOS (days) (mean ± SD)</td>
<td>3.0 ± 3.0</td>
<td>4.0 ± 3.2</td>
</tr>
<tr>
<td></td>
<td>Hospital LOS (days) (mean ± SD)</td>
<td>9.3 ± 25.2</td>
<td>9.2 ± 8.7</td>
</tr>
</tbody>
</table>

*P < 0.05 by t-test or chi-square. ASD, atrial septal defect; ↓ BP, decreased blood pressure; CPB, cardiopulmonary bypass; CPR, cardiopulmonary resuscitation; ECMO, extracorporeal membrane oxygenation; IABP, intra-aortic balloon pump; ICU, intensive care unit; LOS, length of stay; SD, standard deviation; VAD, ventricular assist device; VSD, ventricular septal defect. Reproduced with permission from Andropoulos DB, Stayner SA, Skjonsby BS et al. Anesthetic and perioperative outcome of teenagers and adults with congenital heart disease. J Cardiothorac Vasc Anesth 2002; 16: 731–6.

1. Establish large bore intravenous access and provisions for rapid infusion of volume. A pressurized rapid infusion system capable of delivering at least 500 mL/min of warmed fluid or blood is recommended. In the case of massive bleeding, rapid infusion can be established utilizing the bypass machine. Tubing from the venous reservoir is passed through a roller pump head and connected to large bore venous access. The patient is heparinized, and large volumes can be transfused while preparations are made to rapidly institute bypass via the femoral route.

2. Multifunction external pacing, defibrillating, and cardioversion pads should be applied and antiarrhythmic drugs should be available. The patient’s cardiac rhythm should be assessed, particularly the functioning of pacemakers and underlying cardiac rhythm in case of pacemaker failure.

3. Anesthetic planning should include a detailed preoperative assessment of the patient’s unique pathophysiology and anticipated response to anesthetic interventions should be developed. This is particularly important for the single ventricle patient with poor ventricular function, who may be int tolerant to myocardial depressants, positive pressure ventilation, or loss of sinus rhythm.

General operating room care:

1. Establish large bore intravenous access and provisions for rapid infusion of volume. A pressurized rapid infusion system capable of delivering at least 500 mL/min of warmed fluid or blood is recommended. In the case of massive bleeding, rapid infusion can be established utilizing the bypass machine. Tubing from the venous reservoir is passed through a roller pump head and connected to large bore venous access. The patient is heparinized, and large volumes can be transfused while preparations are made to rapidly institute bypass via the femoral route.

2. Multifunction external pacing, defibrillating, and cardioversion pads should be applied and antiarrhythmic drugs should be available. The patient’s cardiac rhythm should be assessed, particularly the functioning of pacemakers and underlying cardiac rhythm in case of pacemaker failure.
immediately available. In pacemaker-dependent patients who have very slow or non-existent underlying ventricular escape rhythms, a preoperative transvenous pacemaker should be considered.

3 A preoperative discussion between the surgeon, anesthesiologists, and perfusionist should include plans for emergency femoral bypass if necessary.

4 Preparations should be made to treat postoperative hemorrhage. Tranexamic acid, E-aminocaproic acid, and aprotinin are effective in reducing bleeding in these patients. Adequate blood products, including platelets, fresh frozen plasma, and cryoprecipitate should be available. Cell salvage, with reinfusion of washed autologous red blood cells, is appropriate. Thromboelastography during bypass, with heparinase and celite added to neutralize heparin and speed results, may be particularly useful to predict the need for blood products post-bypass, particularly in patients with baseline coagulopathy of cyanosis.

5 Transesophageal echocardiography is indicated for congenital heart surgery in infants and children; these guidelines are also applicable to adult congenital heart surgery.

6 Neurologic monitoring with transcranial Doppler ultrasound (to assist in detecting and limiting cerebral emboli), bispectral index, and near-infrared spectroscopy may be helpful in minimizing neurological complications.

Conclusion

As the number of operations for adult CHD increases, these surgeries will be performed in a variety of institutions and systems. The optimal environment for performing congenital heart surgery on adult patients may be lacking in many situations. In our opinion, this type of surgery is best accomplished in a system designed for adults with CHD. Optimal care for these patients is provided by cardiologists trained and experienced in both pediatric and adult cardiology, by surgeons with training and experience with CHD, and by anesthesiologists with interest and experience in caring for the adult with CHD. Whatever the setting, the cardiac anesthesiologists performing these cases must be thoroughly aware of the anesthetic implications for the unique pathophysiology of each patient, and must not rely on their “usual” expectations of either true pediatric CHD or acquired adult heart disease.

Summary of anesthetic issues for congenital heart lesions most commonly encountered in adults

Atrial septal defect:
- Primarily left-to-right shunt, but may have paradoxical emboli.
- Many patients with hemodynamically insignificant ASDs present after embolic stroke.
- Pulmonary vascular disease usually does not develop until 40 years of age.

Ventricular septal defect:
- Left-to-right shunt.
- Delayed closure may leave longstanding ventricular dysfunction or irreversible pulmonary hypertension.
- Increased incidence of aortic insufficiency (AI).

Patent ductus arteriosus:
- Longstanding left-to-right shunt.
- May develop end-stage pulmonary hypertension.
- Ductus may be calcified or aneurysmal when repaired in adulthood.

Coarctation of the aorta:
- Arterial monitoring in right arm.
- Primary repair in adulthood is associated with poor outcome.
- Revision of childhood repair common, either surgically or via cardiac catheterization.

d-Transposition of the great arteries (D-TGA):
- Atrial arrhythmias and/or sick sinus syndrome after Mustard or Senning procedure.
- Progressive right ventricle failure or tricuspid insufficiency may develop after Mustard or Senning procedure:
  - Tricuspid valve repair or replacement;
  - Conversion to the arterial switch procedure may be indicated, usually preceded by pulmonary artery banding.

Congenitally corrected transposition of the great arteries:
- Arrhythmias (heart block) is common.
- Right (systemic) ventricular failure develops with increasing age.
- Double switch procedure places the left ventricle as the systemic pump.

Ebstein’s anomaly:
- Adults may show congestive heart failure (CHF) or cyanosis depending on right ventricular output.
- Atrial arrhythmias are common, and atrioventricular (AV) block is common after tricuspid replacement.

Tetralogy of Fallot:
- Primary repair can be performed in adults with good outcome.
- Reoperation most commonly needed for pulmonary insufficiency or conduit failure.
- Ventricular arrhythmias are common years after repair.

Atrioventricular canal:
- Most frequently associated with Down syndrome.
- Residual or progressive mitral regurgitation may necessitate surgery later in life.

Truncus arteriosus:
- Essentially all patients require repeat operations for right ventricle to pulmonary artery conduit revision.
- Some patients will require truncal (neo-aortic) valve repair or replacement.
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Single ventricle:
- Variable anatomy (usually atresia of AV valve or semilunar valve) with mixing of systemic and pulmonary venous blood.
- Fontan procedure performed as staged surgical repair: Central venous pressure is the driving force for pulmonary blood flow; Positive pressure ventilation will increase intrathoracic pressure and decrease pulmonary blood flow, thereby decreasing cardiac output.
- Conversion of atrophicpulmonary Fontan to extracardiac Fontan has been performed in adults with improvement in cardiac function.

References

CHAPTER 13 Approach to the teenaged and adult patient


46 Abbott ME. Coarctation of the aorta of adult type: II. A statistical study and historical retrospective of 200 recorded cases with autopsy, of stenosis or obliteration of the descending arch in subjects above the age of 2 years. Am Heart J 1928; 3: 392–421.

47 Reifenstein GH, Levine SA, Gross RE. Coarctation of the aorta: A review of 104 autopsied cases of the “adult type,” 2 years of age or older. Am Heart J 1947; 33: 146–68.


PART 3 Preoperative considerations


